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發病率；

黏膜皮膚淋巴結徵候群

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Kawasaki disease in Hong Kong, 1994 to 2000

1994年至2000年香港的川崎氏病

Objective. To describe the epidemiology, clinical characteristics, and management of Kawasaki disease in children in Hong Kong.

Design. Retrospective survey of medical records from July 1994 to June 1997, and prospective data collection from July 1997 to June 2000.

Setting. Hospitals with a paediatric unit in Hong Kong.

Patients. Patients diagnosed with Kawasaki disease between July 1994 and June 2000 in public hospitals in Hong Kong.

Main outcome measures. Incidence of Kawasaki disease and coronary artery aneurysm rates.

Results. A total of 696 cases of Kawasaki disease were reported. There were 435 (62.5%) boys and 261 (37.5%) girls giving a male to female ratio of 1.7:1. The age ranged from 1 month to 15 years 5 months with a median of 1.7 years. Infants (<1 year) constituted the largest group of patients (223, 32.0%) and overall, 638 (91.7%) were younger than 5 years. Skin rash, conjunctivitis, and oral signs were among the principal clinical features present in over 80% of cases. Prominent cervical lymph nodes larger than 1.5 cm were less commonly found (24%). Coronary artery aneurysms or ectasia were present in 15.7% (109/696), 8.5% (59/696), and 5.0% (35/696) of patients at 2, 4, and 8 weeks, respectively. The incidence of Kawasaki disease per 100 000 children under 5 years was significantly higher in the prospective study period than in the retrospective period (39 vs 26, $P<0.001$).

Conclusion. The incidence of Kawasaki disease is high in Hong Kong and is 39 per 100 000 children below 5 years of age. The coronary artery aneurysm prevalence is 5%. Intravenous gamma-globulin and high-dose aspirin is the mainstay of treatment.

目的：描述香港兒童患上川崎氏病的流行病學情況、臨床特徵和處理。

設計：對1994年7月至1997年6月的醫療記錄進行回顧統計，並於1997年7月至2000年6月期間進行前瞻式數據搜集。

安排：香港設有兒科部門的醫院。

患者：1994年7月至2000年6月期間，在香港的公立醫院被診斷患上川崎氏病的病人。

主要結果測量：川崎氏病發病率和冠狀動脈瘤發病率。

結果：在上述調查期內共有696宗病例，男孩435人（佔62.5%），女孩261人（佔37.5%），男女比例為1.7:1。患者年齡介乎1個月至15歲5個月，中位數為1.7歲。病人以嬰兒（不足1歲）佔最多（223人，佔32.0%），而有638人（佔91.7%）接受診斷時不足5歲。超過80%病人有皮膚出疹、結膜炎、牙齒異狀等典型臨床徵狀，但頸部淋巴腺超過1.5 cm的情況則比較少（24%）。出生後2星期、4星期和8星期的嬰兒中，出現冠狀動脈瘤或冠狀動脈擴張的情況分別是15.7%（109人）、8.5%（59人）和5.0%（35人）。每10萬名5歲以下兒童的感染率，在前瞻研究階段明顯高於回顧研究階段（39人對26人， $P<0.001$ ）。

結論：香港的川崎氏病發病率高，每10萬名5歲以下兒童有39人染病；

而出現冠狀動脈瘤的比例為 5%。經皮免疫球蛋白及高劑量阿司匹林是治療的主要方法。

Introduction

The syndrome of Kawasaki disease (KD) was first described by Dr Tomisaku Kawasaki 38 years ago. It was initially termed mucocutaneous lymph node syndrome,¹ and affects mainly children aged 5 years or under. Diagnosis of KD remains an art: there is no single diagnostic confirmatory test for the disease and it can be camouflaged by the presenting features of different viral infections. Diagnosis is thus based on characteristic clinical signs and symptoms that are classified as principal clinical findings. The congenial interpretation of signs by experienced paediatricians with the aid of other clinical and laboratory findings is most important in the recognition of the disease and in the institution of timely treatment. Accurate diagnosis and early therapeutic intervention within 10 days of onset of fever with high-dose aspirin and intravenous γ -globulin (IVGG) can reduce the risk of developing coronary artery abnormalities from 20%-30% to 5%-10%.²⁻⁸ Thrombosis within an aneurysm, myocardial infarction, and death may occur in the acute phase of the illness.⁹ Patients can also suffer long-term morbidity as a result of scarring of coronary arteries with stenosis, intimal thickening, and accelerated atherosclerosis. Kawasaki disease is the leading cause of acquired heart disease in children in Hong Kong.

The incidence of KD varies in different parts of the world although it is most common in Asia. The reported mean annual incidence per 100 000 children below 5 years in Japan,¹⁰ the United States,¹¹⁻¹³ Britain,¹⁴ and Australia¹⁵ were 110, 8.0 to 47.7, 3.6, and 3.7, respectively. A retrospective study of the incidence of KD in Beijing from 1995 to 1999 ranged from 18.2 to 30.6 per 100 000 children below 5 years old.¹⁶ Kawasaki disease was reported in Hong Kong as early as 1985.¹⁷⁻¹⁹ Ten years ago the incidence in Hong Kong, based on cases seen at one public hospital between 1989 and 1994, was estimated to be 25.4 per 100 000 children below 5 years.²⁰ There was no territory-wide study and the Hong Kong Kawasaki Disease Study Group (HKKDSG) was subsequently formed to study the incidence and characteristics of KD in Hong Kong.

Methods

The HKKDSG was formed in 1993 and comprised paediatric cardiologists and paediatricians. A checklist (Appendix 1) and a surveillance form (Appendix

2) with diagnostic criteria and questions on management and clinical progress were designed and a retrospective survey of KD was carried out in 1997. Patients diagnosed with KD between July 1994 and June 1997 were identified from a hospital audit database. Surveillance forms were completed based on information available in their hospital records. From July 1997 to June 2000, prospective data collection was started in all hospitals using the same documentation. Surveillance forms were sent to the group for periodical audit and review. This study summarises the surveillance results of the 1994 to 1997 retrospective study and the prospective data collected from 1997 to 2000.

There is no confirmatory diagnostic test for KD. Diagnosis is based on recognition of five of the six principal clinical features of the illness,²¹ which include persistent fever, and in the absence of any other obvious pathology:

1. Fever persisting for at least 5 days (many experts believe that when the classic features are present, KD can be diagnosed by experienced observers before day 5);
2. Changes in extremities: acute phase—erythema and oedema of the hands and feet; convalescent phase—membranous desquamation of the fingertips;
3. Polymorphous exanthema;
4. Bilateral, painless bulbar conjunctival injection without exudate;
5. Changes in lips and oral cavity: erythema and cracking of lips, strawberry tongue, diffuse injection of oral and pharyngeal mucosae; and
6. Cervical lymphadenopathy (>1.5 cm in diameter).

Atypical or incomplete KD was diagnosed and recorded on the surveillance form when three or four of the principal criteria plus coronary ectasia²² were present. During the prospective study period, echocardiography was performed at diagnosis, and in the 2nd, 4th, and 8th week following the first day of fever in compliance with the checklist guidelines.

Population data by age-group and sex for epidemiological interpretation were obtained from the Census and Statistics Department, Hong Kong. The annual incidences of KD were calculated by dividing the number of KD cases registered each year by the population of children under 5 years old. Results were analysed using the Statistical Package for the Social Sciences (Windows version 11.5; SPSS Inc, Chicago [IL], United States). Differences were evaluated with the Chi squared test.

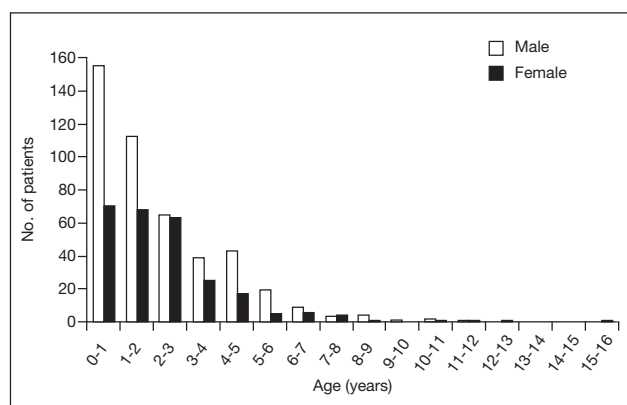
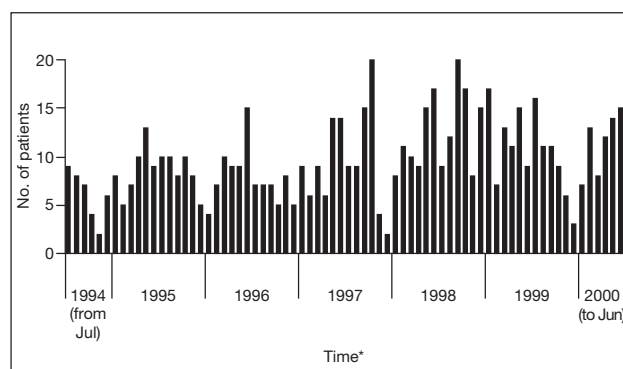


Fig 1. Age and sex distribution of Kawasaki disease in Hong Kong, 1994 to 2000



* Each bar represents the total number of patients with Kawasaki disease in a month

Fig 2. Distribution of Kawasaki disease, 1994 to 2000

Table. Proportion of principal clinical features of patients with Kawasaki disease

Principal clinical feature	No. of patients
Fever	651 (94%)
Rash	599 (86%)
Conjunctivitis	613 (88%)
Oral changes*	598 (86%)
Lip fissuring	393 (56%)
Strawberry tongue	153 (22%)
Oral erythema	264 (38%)
Extremity changes*	504 (72%)
Oedema (hand)	265 (38%)
Erythema (palm)	222 (32%)
Desquamation	206 (30%)
Cervical lymph nodes (>1.5 cm)	170 (24%)

* More than one clinical feature may appear in a patient

Results

Over the 6-year period, a total of 696 cases of KD were reported and only 10 (1.4%) were non-Chinese. There were 435 (62.5%) boys and 261 (37.5%) girls giving a male to female ratio of 1.7:1. The age range was 1 month to 15 years 5 months with a median of 1.7 years. Of these 696 cases, 638 (91.7%) were below 5 years; 223 (32.0%) were younger than 1 year; 415 (59.6%) were 1 to 5 years old; and 58 (8.3%) were older than 5 years. Infants formed the largest group. The age and sex distribution is shown in Fig 1. The frequency of the presenting signs and symptoms is listed in the Table. Skin rash, conjunctivitis, and oral signs were present in over 80% of cases whereas large cervical lymph nodes were less common (24%). The presentation was 'atypical' in 72 (10.3%) patients, 36% of whom were infants. A significantly higher percentage of infants presented with atypical features compared with children aged 1 to 5 years (21% vs 6%, $P<0.001$). There were only six (1%) recurrences within 1 year.

Intravenous γ -globulin was administered as treatment to 589 (84.6%) children. The starting dose was 1 g/kg in 146 (25%) patients and 2 g/kg in 443 (75%) patients. Additional doses were given to 52 (7.5%) patients with persistent or recurrent fever. Persistent fever was defined as fever continuing 48 hours after commencement of IVGG. Fever subsided within 12 hours in one third of patients receiving either IVGG regimen although significantly more patients remained febrile after 48 hours in the 1 g/kg group (24% vs 10%, $P<0.001$). The prevalence of coronary abnormalities at week 8 after onset of disease was also higher in the 1 g/kg group (11.6% vs 3.5%, $P<0.0005$).

The occurrence of KD was significantly higher in the prospective study period than the retrospective period (39 vs 26 per 100 000 children under the age of 5 years, $P<0.001$).

Among all patients, coronary artery aneurysms (CAA) or ectasia were present in 109 (15.7%), 59 (8.5%), and 35 (5.0%) patients in the 2nd, 4th, and 8th week, respectively. Giant aneurysms (diameter ≥ 8 mm) were present in three (0.4%) cases. There were no deaths.

The occurrence of KD was spread evenly over the 6-year period (Fig 2) with peaks occurring in late spring and summer and troughs in November to December (late autumn and early winter when it is dry and cool in Hong Kong).

Discussion

This is the first territory-wide epidemiological study of KD in Hong Kong that compared retrospectively and prospectively collected data. The study included all cases from the public hospitals. Most patients

diagnosed in the private sector with KD were referred to a public hospital for evaluation, treatment, and follow-up because of the high cost of echocardiography and IVGG. Nonetheless it is possible that a few cases may not have been reported, thus the true incidence of KD in Hong Kong may be higher than the 39 per 100 000 children below 5 years old reported here. An incidence of 18.2 to 30.6 per 100 000 children younger than 5 years has been reported in Beijing¹⁶ and 54.9 per 100 000 children younger than 5 years in 1998 in Taiwan.²³ This suggests that the incidence of KD in Chinese population is approximately one third to half of the Japanese and much higher than in Caucasians.

The incidence of KD obtained in the two study periods was significantly different (39 vs 26 per 100 000 children under the age of 5 years for prospective and retrospective study, respectively). This may be due to different study methodologies, a genuine increased incidence in the prospective study period, or, more likely, an increased awareness of the disease by primary physicians, paediatricians, and cardiologists. Low reporting is the common reason for a low incidence in the early years of reports and a subsequent increased incidence equates to increased awareness.

In this study the occurrence of KD peaked in the spring and summer and was lower in autumn and early winter (November to December). This is in agreement with other series¹⁰ and with that reported in China.¹⁶

As in other series, the presence of lymph nodes as a presenting feature was less frequent than the other principal features. The number of children who presented with cervical lymph nodes was fairly low and cervical lymph nodes larger than 1.5 cm in size were only present in 24%. Notably, more children had multiple cervical lymph nodes ($n=308$, 44%). It is not known whether this is associated with clinical characteristics. The Diagnostic Guidelines of Kawasaki Disease produced in 2002 by the Japan Kawasaki Disease Research Committee²⁴ has modified the statement that the sixth principal criteria is "acute non-purulent cervical lymphadenopathy" and no longer states a required size.

Coronary artery aneurysms or ectasia were present in the 4th week in 8.5% (59/696) of children and in 5.0% at 8 weeks. This may be because some investigators used a protocol that required echocardiography to be performed at 2 and 8 weeks only. The checklist

used in this study recommended echocardiography at week 4. This might have increased the number of cases of ectasia and CAA (8.5%) diagnosed at 4 weeks, some of which had resolved by the 8th week (5.0%). This is in accordance with the findings of a Japanese study that reported no new lesions detected by echocardiography between day 30 and 60 in 85 cases of KD.²⁵ Another study of 91 patients in the United Kingdom⁸ reported a maximum incidence of coronary arterial lesions in the 4th week. It is important to perform echocardiography in the 4th week: there are cases of transient dilatations in the early stages at diagnosis and at 2 weeks that account for the higher incidence of CAA in the acute stage that peaks at around 4 weeks and gradually resolves at 8 weeks in the convalescent stage. The American Heart Association (AHA) recommendation in 2004²⁶ to perform echocardiography at 2, 6, and 8 weeks for uncomplicated cases of KD is similar to our practice. Long-term echocardiographic follow-up of patients in this study revealed further resolution of CAA or ectasia in 6 months and up to 2 years.

Atypical KD or 'incomplete KD' accounted for 10% of patients. Such patients often present with persistent fever but incomplete features of three or fewer principal criteria. The diagnosis is often difficult and consultation with an expert is important. Additional laboratory criteria may help confirm a diagnosis (serum albumin, ≤ 3.0 g/L; anaemia for age; elevated alanine aminotransferase; high platelet count after 7 days, $\geq 450\ 000$ /mm³; white blood cell [WBC] count, $\geq 15\ 000$ /mm³; and urine ≥ 10 WBC per high power field). The 2004 AHA statement has proposed a clinical algorithm that is most helpful for the evaluation and treatment of 'incomplete KD'.²⁶

In the absence of specific diagnostic criteria for KD, other diseases with similar clinical features should be excluded (Box).²⁶ Despite this, patients previously diagnosed with measles, viral infections, or scarlet fever have subsequently been found to have KD. Therefore a high index of suspicion must be maintained in a febrile child with rash.

Conclusion

Kawasaki disease is common in Hong Kong Chinese children. This prospective study shows that the incidence is 39 per 100 000 children below 5 years, one third that of Japan. Intravenous γ -globulin together with high-dose aspirin is the mainstay of treatment. The CAA prevalence is 8.5% at 4 weeks and 5.0% at 8 weeks even with IVGG treatment. Additional

Differential diagnosis of Kawasaki disease—diseases and disorders with similar clinical findings

Viral infections (eg measles, adenovirus, enterovirus, Epstein-Barr virus)
 Scarlet fever
 Staphylococcal scalded skin syndrome
 Toxic shock syndrome
 Bacterial cervical lymphadenitis
 Drug hypersensitivity reactions
 Stevens-Johnson syndromes
 Juvenile rheumatoid arthritis
 Rocky mountain spotted fever
 Leptospirosis
 Mercury hypersensitivity reaction (acrodynia)

echocardiography should be performed at 4 weeks to detect CAA or ectasia during the subacute stage of the disease. Atypical KD occurs mainly in infants and accounted for 10% of all cases.

Appendices

Additional material related to this article can be found on the HKMJ website. Please go to <<http://www.hkmj.org.hk>>, search for the appropriate article, and click on Full Article in PDF following the title.

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Appendix 1. Checklist for Kawasaki disease

Checklist for Kawasaki disease Hong Kong Kawasaki Disease Study Group		Hospital No.: Name: I.D. No.: Sex: Age: Team: Ward: Bed:			
I. Clinical findings		Onset		Disappearance	
A. 1. Fever persisting for more than 5 days					
2. Non-purulent conjunctival injection					
3. Mouth changes:					
(a) erythema of oral and pharyngeal cavity					
(b) fissuring of lips					
(c) strawberry tongue					
4. Extremity changes:					
(a) oedema of hand / feet					
(b) erythema of palm / sole					
(c) desquamation of finger tips and toes					
5. Exanthem (polymorphous, macular, non-bullous, non-crusting) spreading from extremity to trunk					
6. Lymphadenopathy					
B. 1. Vomiting / diarrhoea					
2. Arthritis / arthralgia					
3. Tachycardia / CHF					
4. Obstructive jaundice					
5. Aseptic meningitis					
6. Pyuria / urethritis					
7. Others					
II. Laboratory data		Date:			
ESR*					
WBC					
Platelet					
Hb					
Albumin					
C-reactive protein*					
ALT					
Monospot*					
Throat swab*					
Blood culture					
ASOT*					
Virus Study* throat swab, rectal swab					
* optional					
III. Ultrasound findings		Date:			
Coronary artery aneurysms		at diagnosis		2 wks after onset	
LCA (in mm)				4 wks	
RCA (in mm)				8 wks	
Pericardial effusion					
Hydrops of gall bladder					
Others					
IV. Treatment		Started on		Ended on	
Aspirin 30 mg/kg/day					
3-5 mg/kg om					
IVGG 2 g/kg I.V. over 10-12 hours					
or 1 g/kg I.V. over 6 hours					
Others *					
(i) Blood pressure measured (may have hypertension)					
(ii) Palpate all peripheral arteries for obvious aneurysms					
(iii) ECG – on admission					
– at discharge					
– at 2 months after discharge					
(iv) CBP, platelet on admission, at discharge, every 2 weeks until normal					
(v) 2-dimensional echocardiography (2DE) to be done on:					
Day 1 of diagnosis					
2nd, 4th weeks and 8 weeks (if abnormal), then periodically until normal					
(vi) Treatment:					
(1) Initial: anti-inflammatory dose, aspirin 30 mg/kg/day in four divided doses					
(2) Change to 3-5 mg/kg/day in one dose after the acute phase					
(3) Maintain on this dose for 2 months or until platelet/aneurysms have returned to normal					
V. OPD follow-up					
Should not receive MMR and chicken pox vaccine until ≥6 months later.					
Long-term follow-up – for those who have had aneurysm.					
Coronary angiography for those who had giant aneurysm formation or symptoms/signs of myocardial ischaemia.					

Appendix 2. Surveillance form

Surveillance of Kawasaki disease in Hong Kong

(Hong Kong Kawasaki Disease Study Group)

1. Name of patient: (initials e.g. CKM for Chan Ka Mei) _____
2. Sex: _____ 3. DOB (ddmmyy): _____
4. Race: ☐ Chinese (C) ☐ non-Chinese (NC)
5. District (e.g. Shatin, Mongkok, Wanchai, etc.): _____
6. Name of Doctor i/c: _____ Tel of Doctor i/c: _____
7. Name of Hospital: _____ Hospital No.: _____
8. Date of fever onset (ddmmyy): _____
9. Diagnostic criteria Please tick (description if any)

1. Fever >5 days	<input type="checkbox"/>	_____
2. Rash	<input type="checkbox"/>	_____
3. Conjunctivitis	<input type="checkbox"/>	_____
4. Mouth changes	<input type="checkbox"/>	fissured/bleeding lips
	<input type="checkbox"/>	strawberry tongue
	<input type="checkbox"/>	erythema of oral or pharyngeal cavity
5. Extremity changes	<input type="checkbox"/>	oedema of hand/feet
	<input type="checkbox"/>	erythema of palm/sole
	<input type="checkbox"/>	desquamation of finger tips and toes
6. Lymphadenopathy	<input type="checkbox"/>	Cx lymph node >1.5 cm
	<input type="checkbox"/>	multiple Cx Lns
10. Other significant presenting features: _____
11. Coronary artery aneurysms (infants >2.0 mm; beyond infancy >3.0 mm):

<input type="checkbox"/> Yes with <input type="checkbox"/> ectasia <input type="checkbox"/> aneurysm ;	<input type="checkbox"/> No
At 2 wks after onset: <input type="checkbox"/> Yes RCA size_____mm; LCA size_____mm	<input type="checkbox"/> No
At 4 wks after onset: <input type="checkbox"/> Yes RCA size_____mm; LCA size_____mm	<input type="checkbox"/> No
At 8 wks after onset: <input type="checkbox"/> Yes RCA size_____mm; LCA size_____mm	<input type="checkbox"/> No
12. IVGG:

<input type="checkbox"/> given <input type="checkbox"/> not given <input type="checkbox"/> <D10 <input type="checkbox"/> >D10 (Day_____)
<input type="checkbox"/> 2 g/kg iv infusion <input type="checkbox"/> 1 g/kg iv infusion
<input type="checkbox"/> 400 mg/kg/dose x 5 days iv infusion <input type="checkbox"/> others _____
13. How soon to become afebrile after starting IVGG? ☐ <12 hr ☐ <24 hr ☐ <48 hr ☐ >48 hr

Please fax this sheet to HKKDSG, Fax: 2384 5204; Tel: 2958 6741.

From HKKDSG, Convenor: Dr Ng Yin Ming

Hon. Secretary: Prof Rita Sung